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Case Report

Kikuchi–Fujimoto disease: The great imitator

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Abstract

Kikuchi–Fujimoto disease (KFD) is an unusual clinical entity among tropical countries like Srilanka. It follows the self-limiting course with the background of uncertain pathogenesis Here we report a case of a 15-year-old boy who presented with fever with generalized lymphadinitis. The clinical and histological features overlap with infective, autoimmune, and malignant diseases. It slowly resolves the nature and limited exposure to KFD, posing a considerable challenge.

Keywords

Lymphadenitis, Kikuchi disease, necrotizing lymphadenitis

Introduction

KFD or idiopathic histiocytic necrotizing lymphadenitis is an unfamiliar disease. It was first described in 1972 from Japan (3). It is the higher prevalence among Japanese and other Asian people. It is a rare cause of fever with lymphadenitis in children and young females, which mimics lymphoma, SLE, syphilis, infectious mononucleosis, and Tuberculous lymphadenitis (4). A female predilection is reported (female to male ratio of 4:1) (5). Adults younger than 40yrs are often affected, but KFD is seldom reported in children with a low recurrence rate(5).

Case report

A 15-years-old boy presented with a 3-week history of fever with severe constitutional symptoms. He had symmetrically small and large joint pain simultaneously without early morning stiffness for one week. He denies respiratory symptoms, and there was no contact history or past history of TB. Cardiovascular and genitourinary system reviews were not significant. He had not developed any rashes, oral ulcers, or pruritus.

He was mildly pale, with no icterus and no redness of the eye. He had generalized lymphadenopathy (cervical, axillary, and inguinal) with the largest LN in the R/posterior cervical triangle. His Lymph nodes were non-tender and rubbery consistency. His abdomen was soft and had no clinically palpable organomegaly. Table 1shows the investigations performed.

Table 1: Summary of investigations performed

WBC	5,890 (N-66%, L-30%)
Hb	10.6 g/dl (MCV-83.2, MCHC-32)
PLT	321,000
CRP	31IU/l
ESR	112
Blood culture	No growth
LDH	882
Blood picture	Normocytic normochromic anaemia with marked rouleaux formation Compatible with anaemia of chronic disease
ANA	Negative
C Xray	No significant abnormality detected
Mantoux Test	Negative
Fibrinogen level	2.3g/l (1.8-3.6)
USS Abdomen	Multiple cervical LN-(R&L-L1b, LII, LIII) with increased vascularity and no necrotic area seen. Multiple lymph nodes were seen in the axillary, Inguinal, mesenteric, and para-aortic area Thyroid –normal Spleen -9.4cm
R/Cervical LN biopsy	The fragments of LN show multiple circumscribed foci of necrosis and karyorrhectic debris. Neutrophils and atypical cells are seen. Preserved areas show reactive lymphoid follicles with prominent germinal centers. The immunohistochemical assessment was done with CD3, and CD20 showed a reactive pattern of staining. Features are in keeping with Kikuchi's necrotizing lymphadenitis *(Figure 1)*

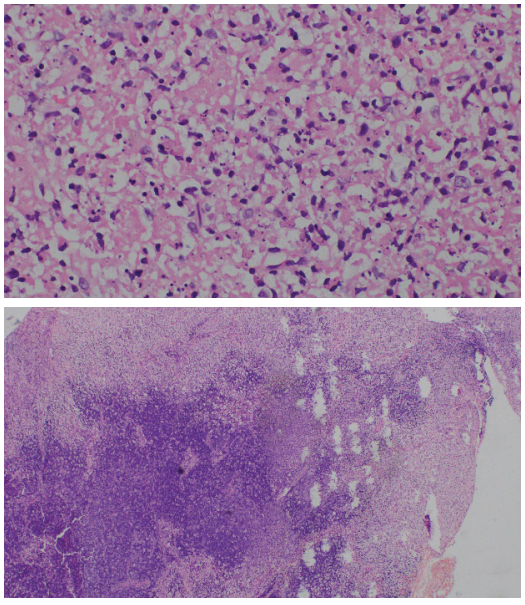

Figure 1: Lymphnode biopsy shows necrotizing lymphadenitis

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Discussion

The clinical manifestations of KFD are fever, general or local lymphadenopathy, skin rash, loss of appetite, weight loss, arthritis, and hepatosplenomegaly. Blood investigation is usually unremarkable and carries less diagnostic value to diagnose KFD (4). The immunohistology findings help to make a definite diagnosis. Histology finding describes three distinct phases as the disease progresses (2). The early stage is a proliferative phase which shows follicular hyperplasia. The next stage is a necrotizing phase in which necrosis with histiocytes as the significant cell type and the absence of neutrophils help differentiate KFD from SLE and drug induce lymphadenopathy. Immunohistochemical stains show CD68-positive plasmacytoid monocytes and histiocytes with an expression of myeloperoxidase help to differentiate from SLE(1). The later stage of the disease shows Xanthomatous appearance in histology.

Histology of Lymph nodes in Hodgkin lymphoma shows necrosis associated with neutrophils infiltrating and reed Sternberg variant atypical cells with CD 15, CD30, and CD 45 positive is easily differentiated from Kikuchi (4) . In herpes simplex, lymph nodes surrounding mononuclear cells and neutrophils are usually present.

KFD has a low recurrence rate of around 3 to 4% (5). Treatment approach usually symptomatic, but persistent or severe symptoms and signs have been treated with glucocorticoids and IV immunoglobulin. Hydroxychloroquine has a place in the management of recurrence of the disease. Few patients may develop SLE in later years, which requires regular follow-up for several years (6). Our patient responded well to symptom management within one month without unnecessary intervention.

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